

Direct effects of mitochondrial dysfunction on poor bone health in Leigh syndrome

韓, 旭

<https://hdl.handle.net/2324/4110461>

出版情報 : Kyushu University, 2020, 博士 (歯学), 課程博士
バージョン :
権利関係 : (C) 2017 Elsevier Inc. All rights reserved.

氏名	韓旭			
論文名	Direct effects of mitochondrial dysfunction on poor bone health in Leigh syndrome (リー症候群における骨障害に対するミトコンドリア機能障害の直接的な影響)			
論文調査委員	主査	九州大学	教授	清島保
	副査	九州大学	教授	久木田敏夫
	副査	九州大学	教授	自見英治郎

論文審査の結果の要旨

Mitochondrial diseases are the result of aberrant mitochondrial function caused by mutations in either nuclear or mitochondrial DNA. Poor bone health has recently been suggested as a symptom of mitochondrial diseases; however, a direct link between decreased mitochondrial function and poor bone health in mitochondrial disease has not been demonstrated. In this study, stem cells from human exfoliated deciduous teeth (SHED) were isolated from a child with Leigh syndrome (LS), a mitochondrial disease, and the effects of decreased mitochondrial function on poor bone health were analyzed. Compared with control SHED, LS SHED displayed decreased osteoblastic differentiation and calcium mineralization. The intracellular and mitochondrial calcium levels were lower in LS SHED than in control SHED. Furthermore, the mitochondrial activity of LS SHED was decreased compared with control SHED both with and without osteoblastic differentiation. Our results indicate that decreased osteoblast differentiation potential and osteoblast function contribute to poor bone health in mitochondrial diseases.

These observations suggest that the LS patient-derived SHED may contribute to investigation the mechanism underlying the decrease of osteoblast differentiation potential and function associated with poor bone health in mitochondrial diseases. Based on this research, the candidate deserves to be conferred the degree of DOCTOR OF PHILOSOPHY (Dental Science) in the Graduate School of Dentistry, Kyushu University.